

Hypoplastic left heart syndrome with right aortic arch in a newborn

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Abstract

A patient with the rare combination of a right-sided aortic arch in hypoplastic left heart syndrome with an incomplete vascular ring is described.

(Br Heart J 1993;69:449-450)

Patients with the usual form of hypoplastic left heart syndrome (HLHS) rarely have a right-sided aortic arch. Not one case was found in a necropsy study of over 200 patients with HLHS.¹ We describe a newborn infant with both abnormalities. These were confirmed by cineangiography and at necropsy.

Case report

A one day old boy, born at term and weighing 2.7 kg, was referred with an imperforate anus, thoracic hemivertebra at T8, hypoplasia of the left thumb, genital anomaly, and a heart murmur over the left sternal border (VATER association). Chromosome study showed a normal male karyotype. Colostomy was performed 2 days later. Heart failure developed at 9 days of age. At presentation the heart rate was 150 beats/min and the respiratory rate was 55/min; the blood pressure in the four limbs was right arm 80/43 mm Hg, right leg 77/31 mm Hg, left arm 74/38 mm Hg, left leg 95/39 mm Hg. A grade 3 systolic murmur was audible over the left sternal border. The lips were dusky and blood gas analysis showed pH 7.282, PCO_2 50.6 mm Hg, PO_2 51.4 mm Hg, HCO_3 24.0 mEq/l, base excess -2.9 mEq/l in room air.

Echocardiography showed situs solitus, mitral stenosis, aortic atresia, a hypoplastic left ventricle, a perimembranous ventricular septal defect, a hypoplastic ascending aorta, and a large duct connecting the main pulmonary trunk and descending aorta. Electrocardiography showed right axis deviation. Chest x ray showed cardiomegaly and increased pulmonary flow. Cardiac catheterisation confirmed the echocardiographic findings. Cineangiography showed HLHS with a right aortic arch. Balloon atrioseptostomy was performed. The patient died of intractable heart failure and metabolic acidosis at 22 days of age.

Postmortem examination showed situs

solitus, drainage of the right superior vena cava and the right inferior vena cava into a right-sided morphologically right atrium with a well-developed eustachian valve. Balloon atrioseptostomy had created an interatrial communication (10 mm in diameter) through the caudal part of a thin-walled septum primum. The superior attachment of septum primum was displaced leftwards in relation to the septum secundum (superior limbus of oval fossa).² Pulmonary veins drained into the left-sided morphologically left atrium. The hypertrophic left atrium was connected to a hypoplastic but hypertrophic morphologically left ventricle through a stenotic and hypoplastic mitral valve. The dimensions of the left ventricular cavity were $2 \times 1 \times 0.5$ cm. A tiny chorda from anterolateral papillary muscle was attached to the apical trabecular part of the ventricular septum. There was a ventricular septal defect (4 mm in diameter) in the perimembranous trabecular portion. Aortic atresia was evident (the external diameter of the ascending aorta was 2 mm). The aorta was connected to the left and right coronary arteries. The external diameter of the right-sided aortic arch was 6 mm, after the left common carotid artery and 5 mm between right subclavian artery and right common carotid artery. A huge pulmonary trunk, 10 mm in external diameter, arose from morphologically right ventricle and was connected to the left-sided patent ductus arteriosus and then to the descending aorta (figure). The aberrant left subclavian artery arose from the descending aorta. The left patent ductus arteriosus and descending aorta had external diameters of 8 mm. The right aortic arch, main pulmonary trunk, and ductus arteriosus formed an incomplete vascular ring, which caused a compression notch at the posterior aspect of the oesophagus. The cardiac apex was to the left.

Discussion

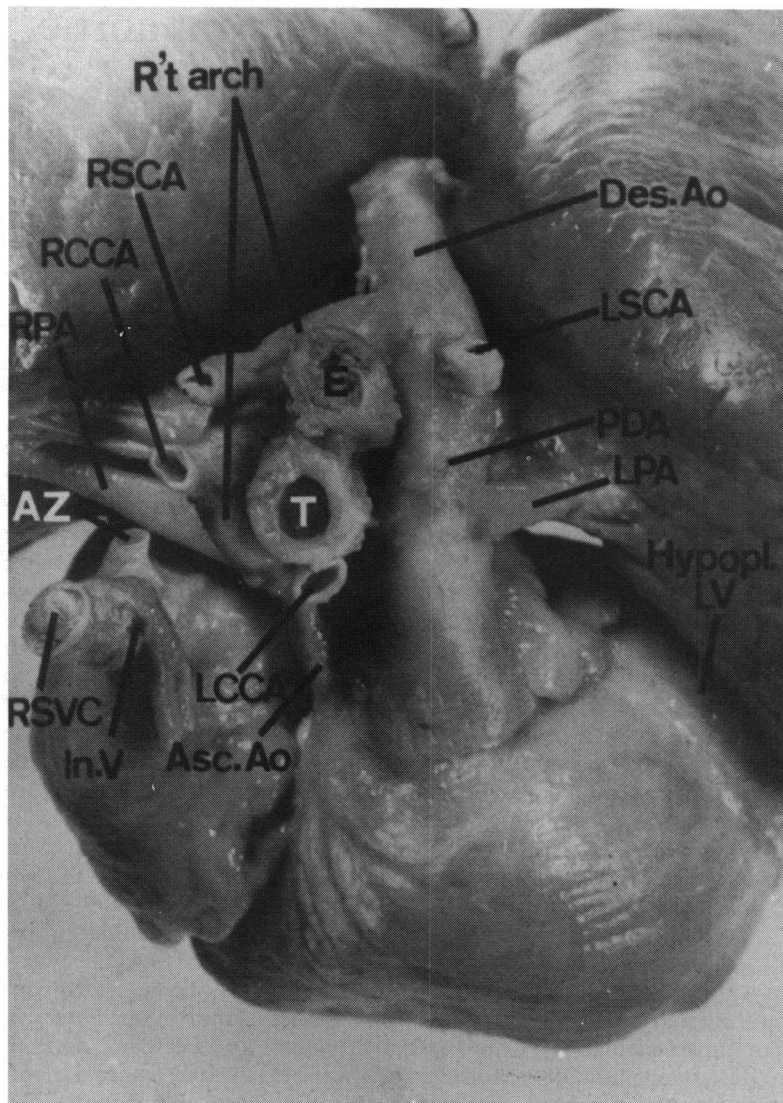
HLHS is a rare cardiac manifestation of VATER syndrome. Until 1990 only two cases of HLHS associated with VATER syndrome had been reported.³ Though our patient had neither a tracheoesophageal fistula nor a renal anomaly he met the diagnostic criteria of VATER syndrome.

HLHS is not uncommon and is universally lethal without surgical intervention.⁴ If our

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Anterocranial view of the heart showing the hypoplastic right aortic arch (R't arch), left patent ductus arteriosus (PDA), and the main pulmonary trunk as the components of the incomplete vascular ring. Asc.Ao, ascending aorta; AZ, azygos vein; Des.Ao, descending aorta; E.o, esophagus; Hypopl. LV, hypoplastic left ventricle; In.V, innominate vein; LCCA, left common carotid artery; LPA, left pulmonary artery; LSCA, left subclavian artery; RCCA, right common carotid artery; RPA, right pulmonary artery; RSCA, right subclavian artery; RSVC, right superior vena cava; T, trachea.

patient had survived until surgery, connection of the pulmonary trunk with the descending aorta with a tube graft, division of the duct plus pulmonary artery banding downstream from the proximal anastomosis site of the prosthesis⁵ through left anterolateral thoracotomy probably would have been the easiest palliative treatment. Because the arch was posteriorly located and no coarctation was present we could have modified the usual Norwood procedure.⁴ The aortic arch could have been reconstructed with a prosthesis. The interatrial communication was wide enough for mixing and with prevention of pulmonary vascular obstructive disease by pulmonary artery banding this baby would have been a possible candidate for later Fontan procedure or heart transplantation.

HLHS may be complicated by anomalies of the aortic arch including coarctation and interruption of the aortic arch.^{2,4,6} But a right aortic arch is rare except among patients with viscerotaxial heterotaxy and left atrial isomerism, and has never been reported in those with situs solitus.^{1,4}

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